Clinical features and diagnosis of Churg-Strauss syndrome (allergic granulomatosis and angiitis)

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INTRODUCTION — The Churg-Strauss syndrome (CSS), also called allergic granulomatosis and angiitis, is a multisystem disorder characterized by chronic rhinosinusitis, asthma, and prominent peripheral blood eosinophilia [1-6]. CSS is classified as a vasculitis of the small and medium sized arteries, although the vasculitis is often not apparent in the initial phases of the disease.

The most commonly involved organ is the lung, followed by the skin. CSS, however, can affect any organ system, including the cardiovascular, gastrointestinal, renal, and central nervous systems. Vasculitis of extrapulmonary organs is largely responsible for the morbidity and mortality associated with CSS.

The clinical features and diagnosis of CSS will be reviewed here. The epidemiology, pathogenesis, treatment, and prognosis of this disorder, as well as the approach to patients with vasculitis and/or eosinophilia are discussed separately. (See "Epidemiology, pathogenesis, and pathology of Churg-Strauss syndrome (allergic granulomatosis and angiitis)" and "Treatment and prognosis of Churg-Strauss syndrome (allergic granulomatosis and angiitis)" and "Classification of and approach to the vasculitides in adults" and "Approach to the patient with eosinophilia".)

CLINICAL FEATURES

Phases of disease — The clinical features of the Churg-Strauss syndrome typically develop in several sequential phases, although these phases are not always clearly distinguishable [7,8]:

- Prodromal phase – The prodromal phase occurs among individuals in the second and third decades of life and is characterized by atopic disease, allergic rhinitis, and asthma.
- Eosinophilic phase – Features of the eosinophilic phase include peripheral blood eosinophilia and eosinophilic infiltration of multiple organs, especially the lung and gastrointestinal tract. Almost 40 percent of patients with CSS present with pulmonary opacities, asthma, and peripheral eosinophilia prior to the development of a systemic vasculitis (polyangiitis) [7].
- Vasculitic phase – In the third and fourth decades of life, a life-threatening systemic vasculitis of the medium and small vessels frequently occurs, and is often associated with vascular and extravascular granulomatosis [6]. The vasculitic phase may be heralded by nonspecific constitutional symptoms and signs, especially fever, weight loss, malaise, and lassitude.

Asthma and lung disease — Asthma is the cardinal clinical feature of CSS and is present in more than 95 percent of patients. Asthma usually precedes the vasculitic phase by approximately 8 to 10 years [6,9]. (See 'Phases of disease' above.)

CSS is typically suspected in patients whose asthma is poorly controlled on moderate doses of inhaled glucocorticoids; many patients diagnosed with CSS require frequent or long-term courses of systemic glucocorticoids to control their asthma. As the vasculitic phase begins, asthma severity and the number of exacerbations may increase. Rarely, the asthma symptoms lessen in the early stages of the vasculitic phase.

Prolonged treatment of asthma with glucocorticoid therapy may partially or totally suppress the usual clinical signs of untreated CSS. The disease may therefore not become evident until glucocorticoids are reduced or stopped [10,11].

Other pulmonary findings are reported in 50 to 70 percent and include pulmonary opacities with eosinophilia, pleural effusion (often eosinophilic), nodules that are rarely cavitary, and alveolar hemorrhage [3].

Upper airway and ear disease — Allergic rhinitis is a common finding in CSS [12]. Other forms of involvement of the nose and paranasal sinuses are common and include nasal obstruction, recurrent sinusitis, and nasal polyposis. In a series of 29 patients with CSS, nasal polyposis was detected in 60 percent [13]. Prior to the onset of eosinophilic infiltration of the lung or other organs, the characteristics of these patients overlap considerably with those associated with aspirin-exacerbated respiratory disease. (See "Clinical manifestations, pathophysiology, and diagnosis of chronic rhinosinusitis", section on 'CRS with nasal polyposis' and "Aspirin exacerbated respiratory disease".)

Chronic serous otitis and sensorineural hearing loss are occasionally seen in CSS and likely reflect the severity of rhinosinusitis [14,15]. Necrotizing lesions of the nasopharynx and upper airway are more characteristic of granulomatosis with polyangiitis (Wegener's), and are unusual in CSS. (See "Clinical manifestations and diagnosis of granulomatosis with polyangiitis (Wegener's) and microscopic polyangiitis".)

Skin — Skin involvement is one of the most common features of the vasculitic phase of CSS. Half to two-thirds of patients with CSS have skin lesions, which usually appear as tender subcutaneous nodules on the extensor surfaces of the arm, particularly the elbows, hands, and legs (picture 1) [3,16]. Biopsy of these lesions usually reveals granulomas. The pathology of cutaneous CSS is discussed in greater detail separately. (See "Epidemiology, pathogenesis, and pathology of Churg-Strauss syndrome (allergic granulomatosis and angiitis)", section on 'Pathology'.)

Skin lesions can also appear as palpable purpura, a macular or papular erythematous rash, and hemorrhagic lesions, ranging from petechiae to extensive ecchymosis [17].

Cardiovascular — Cardiac involvement is one of the more serious manifestations of CSS, accounting for approximately one-half of deaths attributable to CSS [18,19]. Clinical manifestations of cardiac involvement include clinical signs of heart failure and cardiac rhythm abnormalities [18]. Patients with cardiac involvement typically have a shorter duration of CSS related symptoms than those without.

In a series of 49 patients with CSS, 22 had evidence of cardiac involvement based on electrocardiogram, echocardiogram, and contrast-enhanced cardiac magnetic resonance imaging (MRI) [18]. Cardiac abnormalities included an abnormal ECG in all patients; valvular insufficiency, pericardial effusion, and heart failure were noted in 73, 50, and 41 percent, respectively. Endomyocardial involvement was found in 12 patients based on cardiac MRI findings of mural thrombus and a positive endomyocardial biopsy [18]. The majority of patients improved with treatment, although 2 of the patients with endomyocardial disease died of

heart failure. Patients with cardiac involvement were less likely to have a positive ANCA and more likely to have higher peripheral blood eosinophil counts than other CSS patients.

The use of various cardiovascular tests to evaluate myocardial CSS and the typical biopsy findings seen on endomyocardial biopsy are discussed separately. (See 'Cardiovascular tests' below and "Epidemiology, pathogenesis, and pathology of Churg-Strauss syndrome (allergic granulomatosis and angiitis)", section on 'Pathology'.)

Neurologic — A peripheral neuropathy, usually mononeuritis multiplex, is seen in up to 75 percent of patients with CSS [3,6,20-22]. Untreated, this may progress to a symmetric or asymmetric polyneuropathy [23]. Severe neuropathic pain may accompany the peripheral neuropathy [22]. A more detailed description of vasculitic neuropathy is presented separately. (See "Clinical manifestations of vasculitic neuropathy".)

In addition, subarachnoid and cerebral hemorrhage, and cerebral infarction are reported, but rare [6,24,25].

Renal — The frequency of renal involvement varies among studies [26,27]. In the largest series of 116 patients with CSS, renal involvement was found in 31 patients (27 percent) [27]. One-half had rapidly progressive or acute renal insufficiency (plasma creatinine concentration >1.4 mg/dL [>124 micromol/L]), while the others had isolated proteinuria or microscopic hematuria. Sixteen patients underwent renal biopsy, which demonstrated necrotizing glomerulonephritis in 11 patients. A positive test for antineutrophil cytoplasmic antibodies (ANCA) was found in all patients with glomerulonephritis, compared with 26 percent of patients without renal involvement. (See 'Antineutrophil cytoplasmic antibodies' below.)

Systemic hypertension affects approximately 10 to 30 percent of patients with CSS and may reflect renal involvement with CSS or be a complication of glucocorticoid therapy [6,7,24].

Gastrointestinal tract — An eosinophilic gastroenteritis, characterized by abdominal pain (59 percent of patients), diarrhea (33 percent), gastrointestinal bleeding (18 percent), and colitis, may precede or coincide with the vasculitic phase of CSS. (See "Eosinophilic gastroenteritis" and 'Phases of disease' above.)

Musculoskeletal — Myalgias, migratory polyarthralgias, and frank arthritis are less common, but may affect 40 to 50 percent of patients in the vasculitic phase of the disorder [6]. (See 'Phases of disease' above.)

Lymphadenopathy — Eosinophilic lymphadenopathy has been noted in 30 to 40 percent of patients [11]. Prominent cervical and axillary lymphadenopathy has been reported with individual lymph nodes up to 3 cm in diameter.

Complications in pregnancy — The fetal death rate may be slightly increased among pregnancies occurring in patients with CSS, although the number of reported pregnancies is low. As an example, one review reported a fetal death rate of approximately 15 percent among 22 pregnancies [3]. The effect of Churg-Strauss vasculitis on the placenta is not known.

EVALUATION AND DIAGNOSIS — The diagnosis of CSS is typically suspected based on the clinical findings (ie, eosinophilia ≥1500/microL, asthma, allergic rhinitis). However, confirming the diagnosis is often difficult because of the following confounding factors:

- Individual manifestations of the syndrome can occur in isolation.
- Lung parenchymal involvement is not universal.
- Some manifestations can exist for many years before additional features become clinically apparent. (See 'Phases of disease' above.)

- Although CSS is classified as a vasculitis, only 40 to 60 percent of patients with CSS have antineutrophil cytoplasmic antibodies (ANCA). In addition, many biopsies do not show a necrotizing vasculitis or granuloma, but rather an apparently nondestructive infiltration of vessel walls by eosinophils [28]. (See "Clinical manifestations and diagnosis of granulomatosis with polyangiitis (Wegener's) and microscopic polyangiitis".)

Laboratory tests — There are no laboratory tests that are specific for CSS, although eosinophilia is characteristic. For patients who have suspected CSS, we typically obtain a complete cell count with differential, a total eosinophil count, and an IgE level. We also usually obtain an ANCA test, although the sensitivity and specificity are low, as noted below.

Eosinophilia — Peripheral blood eosinophilia (usually 5000 to 9000 eosinophils/microL) is the most characteristic finding, although levels over 1500 cells/microL (or greater than 10 percent of the total leukocyte count) should prompt suspicion for CSS [7,29,30]. Eosinophilia, however, is occasionally missed because of rapid spontaneous, or glucocorticoid-induced reductions or fluctuations in eosinophil counts. Tissue eosinophilia can still be found in patients in whom peripheral blood eosinophilia is absent.

Antineutrophil cytoplasmic antibodies — ANCA are found in 40 to 60 percent of patients with CSS [9,20,31-33]. The majority of ANCA-positive CSS patients (70 to 75 percent) have antibodies directed against myeloperoxidase with a perinuclear staining pattern (called MPO-ANCA or P-ANCA) (picture 2) [34-36]. (See "Clinical spectrum of antineutrophil cytoplasmic antibodies".)

CSS disease manifestations may differ between ANCA-positive and ANCA-negative patients, although confirmatory data are needed [8]. In a series of 112 patients with newly diagnosed CSS, a positive ANCA at diagnosis was associated with renal involvement, peripheral neuropathy, and biopsy-proven vasculitis, whereas a negative ANCA was associated with heart disease and fever [33].

Acute phase reactants — Tests of inflammation such as the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are nonspecific and do not differentiate between flares of CSS and infections (eg, sinusitis, pneumonia). Their usefulness in CSS is unclear. (See "Acute phase reactants", section on 'Clinical relevance'.)

Other laboratory tests — For patients with strongly suspected or known CSS, additional tests are performed to assess the extent of disease, including a urinalysis, serum creatinine, and a serum creatine kinase-MB.

Nonspecific laboratory abnormalities that may be observed in CSS include [6,7]:

- Normochromic, normocytic anemia
- Leukocytosis
- An elevated IgE level is common, and may vary with the activity of the vasculitic process
- Hypergammaglobulinemia
- A positive rheumatoid factor at low titer
- Normal or elevated complement (C3, C4, CH50) levels

Imaging — Chest radiographs are typically obtained during a flare of asthma or to evaluate peripheral eosinophilia. Chest radiographic abnormalities in patients with CSS are diverse and include the following (picture 3A-B) [37-40]:

- Transient and patchy opacities (75 percent of patients), without lobar or segmental distribution

- Symmetrical opacities in an axillary and peripheral distribution
- Opacities radiating from the hilum with hilar adenopathy
- Diffuse interstitial or miliary opacities
- Pulmonary hemorrhage causing widespread shadowing
- Bilateral, nodular disease without cavitation
- Hilar adenopathy (infrequent)
- Pleural effusions, which are found in 30 percent of patients, and are usually exudative and eosinophilic [38]

High resolution chest computed tomography (HRCT) is typically obtained to evaluate unexplained dyspnea, an abnormal chest radiograph, or gas transfer abnormalities, such as a low diffusion capacity or low pulse oximetry. Typical HRCT findings in CSS include peribronchial thickening, areas of septal thickening, and widely scattered patchy, indistinct opacities. One series described parenchymal consolidation or ground glass opacification on the CT scans of 10 of 17 patients [40]. These findings are not specific for CSS, but may guide choice of a location for bronchoalveolar lavage or lung biopsy and help in the assessment of the extent of disease. (See "High resolution computed tomography of the lungs".)

A less common HRCT feature is a significant enlargement of the peripheral pulmonary arteries (when compared to the corresponding bronchi) in combination with a stellate and irregular configuration of some pulmonary arteries (referred to as the vasculitis sign) (picture 4) [39].

Cardiac magnetic resonance imaging is discussed below. (See 'Cardiovascular tests' below.)

Pulmonary function tests — Spirometry typically shows variable airflow limitation (obstruction) consistent with asthma [7]. Some patients may have a component of irreversible airflow limitation noted on spirometry or peak expiratory flow. When lung parenchymal involvement occurs, lung volume measurements (eg, total vital capacity and forced vital capacity) may be decreased. In addition, gas transfer abnormalities may be manifest by a low pulse oximetry at rest or with exertion, or by a decrease in the diffusion capacity. (See "Overview of pulmonary function testing in adults" and "Use of pulmonary function testing in the diagnosis of asthma".)

Bronchoalveolar lavage — Bronchoalveolar lavage (BAL) is typically performed in patients with interstitial opacities on radiographic imaging to evaluate for eosinophilia, infection, alveolar hemorrhage, or malignancy. In CSS, BAL typically reveals a high percentage of eosinophils in the lavage fluid (usually greater than 33 percent). However, this finding is not specific for CSS and would only be present in a patient with active pneumonitis (table 1). (See "Causes of pulmonary eosinophilia" and "Role of bronchoalveolar lavage in diagnosis of interstitial lung disease".)

Biopsy — Surgical lung biopsy, although not always available, is the "gold standard" for the diagnosis of CSS [41]. In contrast, transbronchial lung biopsy is generally not helpful. (See "Role of lung biopsy in the diagnosis of interstitial lung disease".)

When either skin disease or peripheral neuropathy is present, biopsy of one of those sites is less invasive and often preferred to a lung biopsy. In one study, 15 of 28 patients with a peripheral neuropathy and CSS had evidence of a necrotizing vasculitis on a peripheral nerve biopsy [23]. (See "Diagnosis and treatment of vasculitic neuropathy".)

The lung biopsy from patients with CSS may show asthmatic bronchitis, eosinophilic pneumonia, extravascular granulomas, or vasculitis (affecting arteries, veins, or capillaries). The histopathologic findings of CSS are discussed in more detail separately. (See "Epidemiology, pathogenesis, and pathology of Churg-

Strauss syndrome (allergic granulomatosis and angiitis)", section on 'Pathology'.)

Cardiovascular tests — The specific indications for obtaining cardiovascular tests such as the electrocardiogram (ECG), echocardiogram, and cardiovascular magnetic resonance imaging (CMR) are unclear. We typically obtain an electrocardiogram and an echocardiogram (looking for wall motion abnormalities, mural thrombi, and valvular thrombi) as part of the initial evaluation of a patient with a diagnosis of CSS.

If abnormalities are noted on these tests, we perform CMR with gadolinium enhancement, if renal function is adequate. Gadolinium enhancement on CMR has been reported to correlate with endomyocardial biopsy evidence of eosinophilic infiltration [42]. Assessment of late images may reveal myocardial necrosis [43]. However, the sensitivity and specificity of CMR for myocardial involvement with CSS is not known. Gadolinium should be used with caution in patients with advanced renal insufficiency because of the risk of inducing nephrogenic systemic fibrosis. (See "Nephrogenic systemic fibrosis/nephrogenic fibrosing dermopathy in advanced renal failure".)

An endomyocardial biopsy is usually obtained when CSS is suspected, noninvasive testing suggests endomyocarditis, and the documentation of cardiac involvement would affect the decision about whether to initiate immunosuppressive therapy. (See "Endomyocardial biopsy" and "Epidemiology, pathogenesis, and pathology of Churg-Strauss syndrome (allergic granulomatosis and angiitis)", section on 'Pathology'.)

DIAGNOSTIC CRITERIA — A number of different sets of criteria for the diagnosis of CSS have been proposed [7,30,35,44]. The classifications used most commonly are the American College of Rheumatology (ACR, preferred) and the Lanham criteria [7,30].

The ACR has established six criteria for the classification of CSS in a patient with documented vasculitis [30]. The presence of four or more of these criteria had a sensitivity of 85 percent and a specificity of 99.7 percent for CSS:

- Asthma (a history of wheezing or the finding of diffuse high pitched wheezes on expiration)
- Greater than 10 percent eosinophils on the differential leukocyte count
- Mononeuropathy (including multiplex) or polyneuropathy
- Migratory or transient pulmonary opacities detected radiographically
- Paranasal sinus abnormality
- Biopsy containing a blood vessel showing the accumulation of eosinophils in extravascular areas (picture 5A-B)

The Lanham criteria include asthma, peak peripheral blood eosinophilia in excess of 1500 cells/microL, and systemic vasculitis involving two or more extra-pulmonary organs [7]. In this classification, all three criteria must be met for a diagnosis of CSS.

DIFFERENTIAL DIAGNOSIS — The main diseases to consider in the differential diagnosis of CSS are aspirin-exacerbated respiratory disease, the eosinophilic pneumonias, allergic bronchopulmonary aspergillosis, the hypereosinophilic syndrome, granulomatosis with polyangiitis (Wegener's), and microscopic polyangiitis. The following observations may help to narrow the differential diagnosis:

- Aspirin-exacerbated respiratory disease — Aspirin exacerbated respiratory disease (AERD) refers to the combination of asthma, chronic rhinosinusitis with nasal polyposis, and reactions to aspirin (acetylsalicylic acid, ASA) and other COX-1 inhibiting nonsteroidal antiinflammatory drugs (NSAIDs) characterized by bronchoconstriction, nasal congestion, and rhinorrhea. These patients may also

have eosinophilia, but do not have eosinophilic pneumonia or the other organ system involvement seen in CSS. However, AERD may evolve into CSS. (See "Aspirin exacerbated respiratory disease".)

- Chronic eosinophilic pneumonia — Chronic eosinophilic pneumonia usually lacks granulomas on biopsy and generally does not involve organs other than the lung [45,46].

  Occasionally, an episode of eosinophilic pneumonia may precede the other manifestations of CSS, making it difficult to differentiate CSS from chronic eosinophilic pneumonia. These patients are monitored closely for development of evidence of additional organ involvement that would secure the diagnosis of CSS. (See 'Phases of disease' above and "Causes of pulmonary eosinophilia", section on 'Chronic eosinophilic pneumonia'.)

- Allergic bronchopulmonary aspergillosis — Allergic bronchopulmonary aspergillosis is another cause of asthma, radiographic pulmonary opacities, and eosinophilia, but does not affect extrapulmonary organs other than the nose and sinuses. (See "Causes of pulmonary eosinophilia".)

- Hypereosinophilic syndrome — While some patients with the hypereosinophilic syndrome (HES) may have a cough, a minority have pulmonary infiltrates, and asthma is rare. When a clear differentiation cannot be made on clinical grounds, molecular testing for the FIP1L1/PDGFRalpha mutation may be helpful as this is suggestive of HES. (See "Clinical manifestations, pathophysiology, and diagnosis of the hypereosinophilic syndromes", section on 'Hematologic evaluation'.)

- Other vasculitides — Granulomatosis with polyangiitis (Wegener's), microscopic polyangiitis, and CSS can all affect the lung, although the degree of eosinophilia and presence of asthma are typical of CSS and not usually seen in the other two. The type of ANCA seen in CSS is more typically anti-myeloperoxidase, whereas in granulomatosis with polyangiitis it is more likely anti-proteinase 3. (See "Classification of and approach to the vasculitides in adults".)

SUMMARY AND RECOMMENDATIONS

- The Churg-Strauss syndrome (CSS), also called allergic granulomatosis and angiitis, is a multisystem disorder characterized by chronic rhinosinusitis, asthma, and prominent peripheral blood eosinophilia (≥ 1500 cells/microL and/or > 10 percent eosinophils on differential leukocyte count). The exact etiology of CSS is unknown. (See 'Introduction' above.)

- Asthma is the cardinal feature of CSS (occurring in more than 95 percent of patients) and usually precedes the vasculitic phase by approximately 8 to 10 years. (See 'Asthma and lung disease' above.)

- Two-thirds of CSS patients have skin involvement ranging from palpable purpura to subcutaneous nodules. Skin biopsy is often helpful for confirming the diagnosis. (See 'Skin' above.)

- Cardiac involvement is one of the more serious manifestations of CSS, accounting for approximately one-half of deaths attributable to CSS. It should be suspected in the presence of refractory dyspnea, clinical evidence of heart failure, or cardiac rhythm abnormalities. (See 'Cardiovascular' above.)

- Most patients with CSS have peripheral blood eosinophilia (typically above 1500/microL, often 5000 to 9000/microL), although this may be obscured by use of systemic glucocorticoids to control asthma. (See 'Eosinophilia' above.)

- Antineutrophil cytoplasmic antibodies (ANCAs) are noted in 40 to 60 percent of CSS patients. The majority of ANCAs associated with CSS are directed against myeloperoxidase with a perinuclear staining pattern (called MPO-ANCA or P-ANCA). (See 'Antineutrophil cytoplasmic antibodies' above.)
- Typical findings on chest high resolution computed tomography (HRCT) include patchy parenchymal consolidation or ground glass opacification; nodules may also be noted. (See 'Imaging' above.)
- The diagnosis of CSS is suggested by the presence of asthma, rhinosinusitis, and eosinophilia and then confirmed by lung biopsy or biopsy of other clinically affected tissues (eg, skin, peripheral nerve). (See 'Evaluation and diagnosis' above.)
- Two sets of diagnostic criteria are commonly used; the American College of Rheumatology (ACR) criteria for the classification of CSS in a patient with documented vasculitis and the Lanham criteria. (See 'Diagnostic criteria' above.)
- The main diseases to consider in the differential diagnosis of CSS are aspirin-exacerbated respiratory disease, the eosinophilic pneumonias, allergic bronchopulmonary aspergillosis, the hypereosinophilic syndrome, granulomatosis with polyangiitis (Wegener's), and microscopic polyangiitis. (See 'Differential diagnosis' above.)

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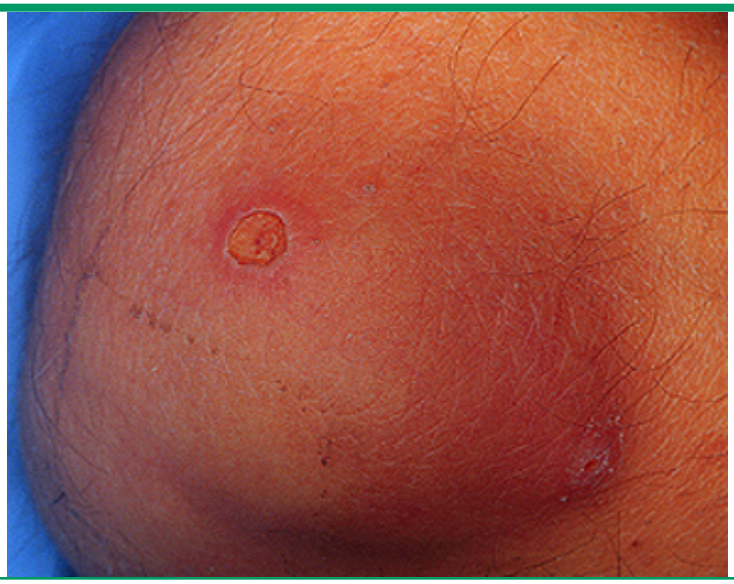
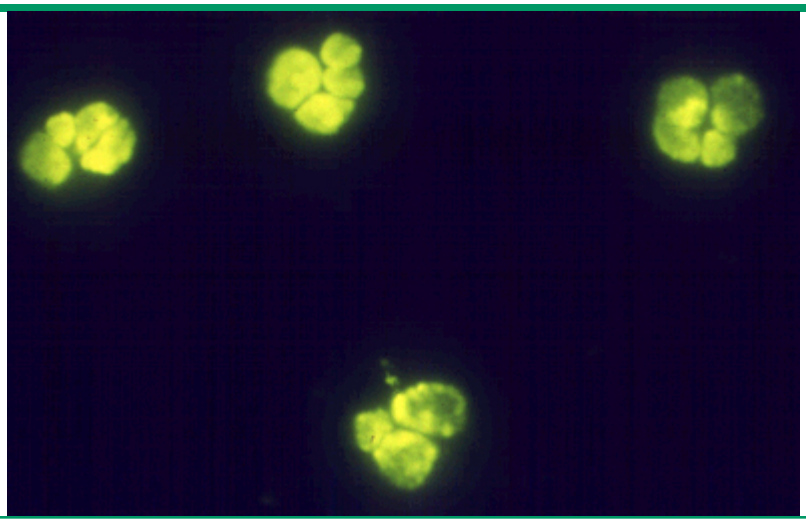
GRAPHICS

Churg-Strauss syndrome



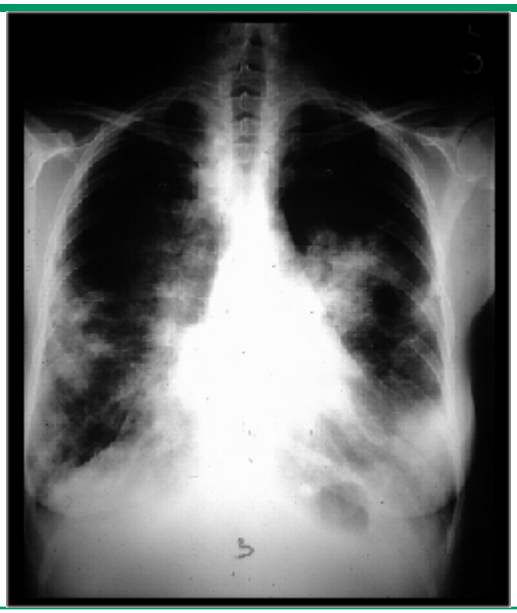
Cutaneous ulceration on the elbow of a patient with the Churg-Strauss syndrome. *Courtesy of Talmadge E King, Jr, MD.*

P-ANCA pattern on indirect immunofluorescence



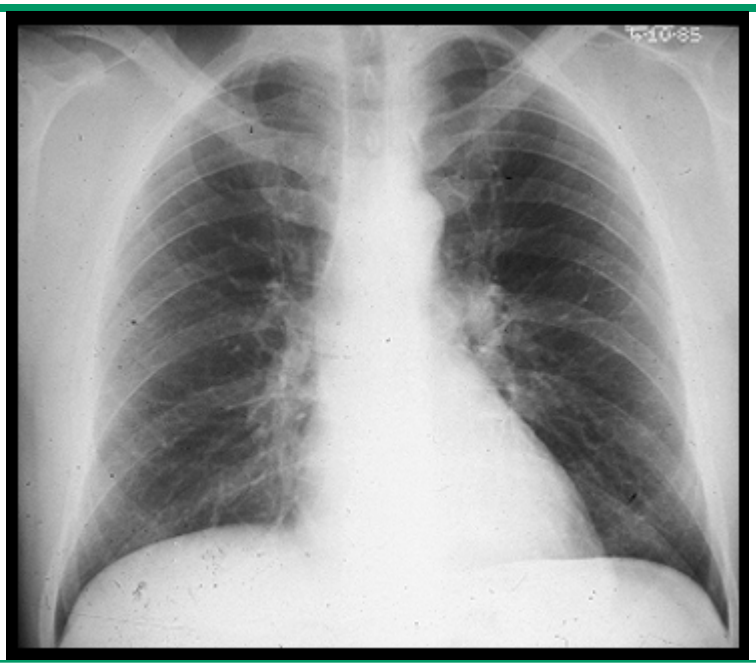
Demonstration of perinuclear antineutrophil cytoplasmic antibodies (P-ANCA) by indirect immunofluorescence with normal neutrophils. Staining is limited to the perinuclear region, and the cytoplasm is nonreactive. Among patients with vasculitis, the antibodies are usually directed against myeloperoxidase. However, a P-ANCA pattern can also be seen with autoantibodies against a number of other antigens including lactoferrin and elastase. Non-MPO P-ANCA can be seen in a variety of nonvasculitic disorders. *Courtesy of Helmut Rennke, MD.*

Churg-Strauss chest radiograph



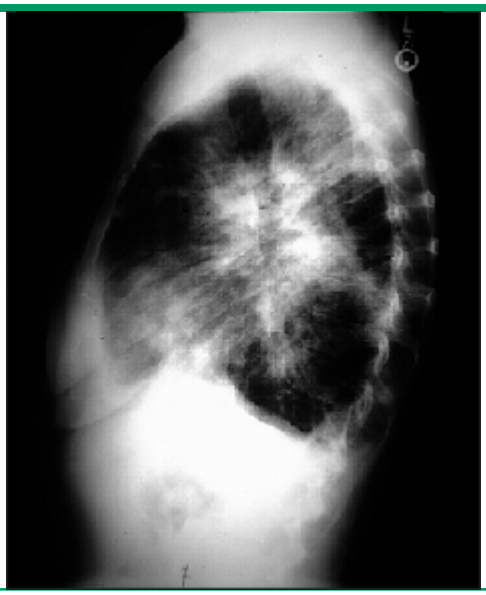
Posteroanterior chest radiograph of a patient with Churg-Strauss syndrome demonstrating patchy bilateral consolidative opacities. *Courtesy of Steven E Weinberger, MD.*

Normal chest radiograph



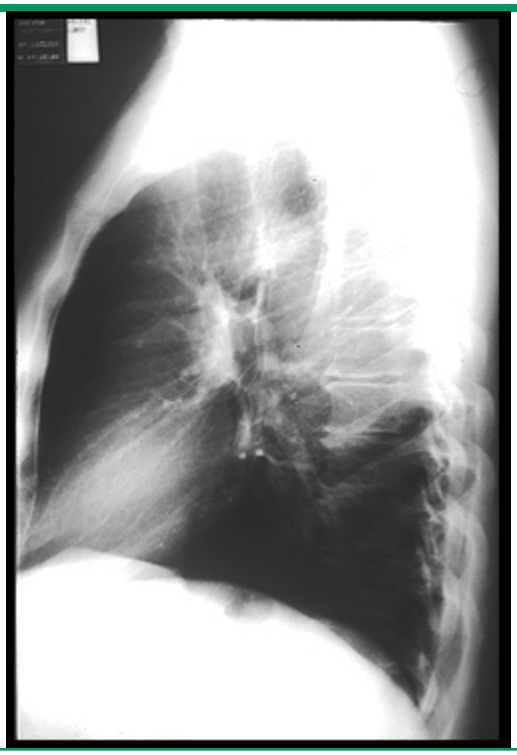
Posteroanterior view of a normal chest radiograph. *Courtesy of Carol M Black, MD.*

Churg-Strauss lateral chest radiograph



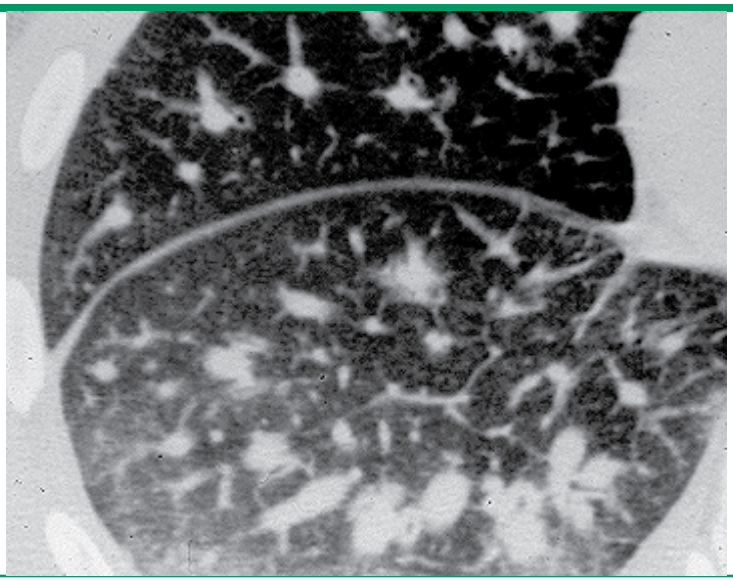
Lateral chest radiograph of a patient with Churg-Strauss syndrome demonstrating prominent consolidative opacities.
Courtesy of Steven E Weinberger, MD.

Normal lateral chest radiograph



Courtesy of Steven Weinberger, MD.

Churg-Strauss syndrome



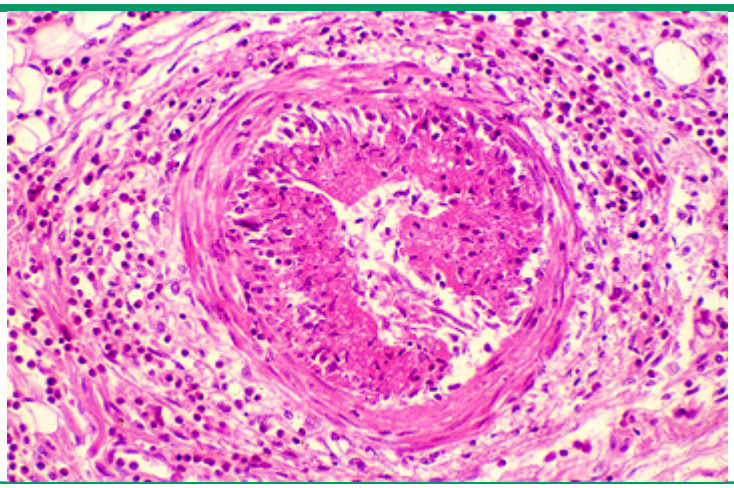
Supine high-resolution CT scan through the right lower and middle lobes in a patient with Churg-Strauss syndrome demonstrates a thickened major fissure and thickened septal lines in the subpleural parenchyma. Enlarged pulmonary arteries are also present in this area. The adjacent bronchi are normal in size with some peribronchiolar thickening. Small patchy opacities are present in the pulmonary parenchyma and are believed to correlate with the eosinophilic pneumonitis that is present. *Courtesy of Talmadge E King, Jr, MD.*

Interstitial lung disease associated with BAL eosinophilia

High count (≥25 percent)
Chronic eosinophilic pneumonia (≥40 percent)
Churg Strauss syndrome with active pneumonitis
Idiopathic acute eosinophilic pneumonia (≥25 percent)
Tropical pulmonary eosinophilia (40 to 70 percent)
Mild to moderate counts (<25 percent)
Connective tissue disease
Drug-induced pneumonitis
Fungal pneumonia
Idiopathic pulmonary fibrosis (<10 percent)
Pulmonary Langerhans cell histiocytosis (Histiocytosis X)
Sarcoidosis

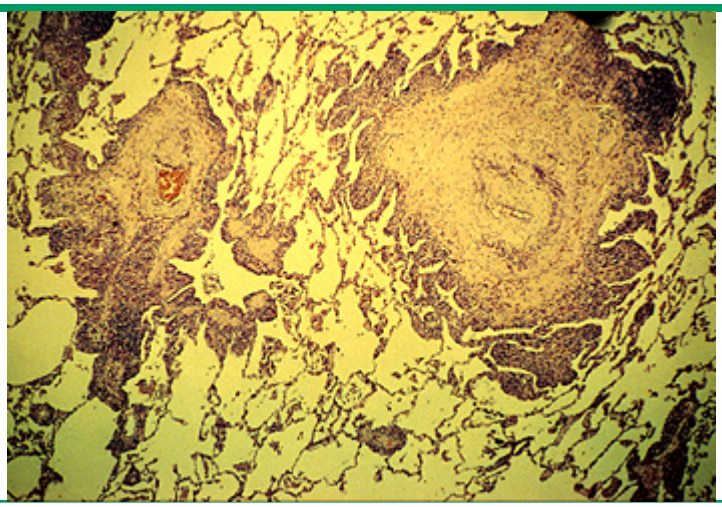
BAL: bronchoalveolar lavage.

Vasculitis and eosinophilic infiltration in Churg Strauss



Small artery in a patient with Churg Strauss syndrome showing intimal fibrinoid necrosis and mural infiltration by histiocytes consistent with a necrotizing granulomatous vasculitis. There is marked extravascular eosinophilia.

Churg-Strauss syndrome



Low power view of a lung biopsy from a patient with the Churg-Strauss syndrome shows well-established, necrotizing and occlusing arterial lesions with obliteration of the arterial lumen. Dilatation of lymphatic channels in the arterial adventitia contributes significantly to the marked size discrepancy between the artery and its adjacent bronchiole. Other regions of the lung reveal marked intraalveolar eosinophilic infiltration in the parenchyma. (Hematoxylin and eosin, x 10). *Courtesy of Talmadge E King, Jr, MD.*